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210 Importance of CF-formula in the nutrition of children with cystic fibrosis

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Milupa Cystilac – cf formula – is a hypercaloric formula based on the specific nutritional needs of children with CF (cystic fibrosis) in their first years of life. Cystilac is higher in energy, fat, protein, vitamins, minerals and trace elements than a formula for healthy children.

The aim of our study was to assess the effectiveness of Cystilac in nutrition for children with CF.

33 children aged from 3 to 15 months were enrolled in this 6 month duration study. 28 children finished the observation. The remaining 5 patients withdrew prematurely because of milk protein allergy. 28 children with CF, which were fed according to their needs with standard formula, were the control group.

Their height, weight and head circumference were measured every 3 months. Blood tests were performed at the beginning and at the end of the study (prealbumins, total protein, sodium, vitamins A and E, RBC, WBC, haemoglobin). Every month acceptance and tolerance of Cystilac were checked. After 2 and 5 months the parents prepared detailed nutritional protocols, analyzed by dietician. Statistical analysis of the studied children and the control group was based on the T-Student test.

There was no statistical difference in somatic development between the groups (short period of observation).

The parameters of vitamin A, E (11.1–17.5 nmol/l), protein (61.4–64.4 g/l), prealbumins (0.243–0.273 g/l) and RBC were however significantly higher in the Cystilac group, as were energy, protein, and carbohydrate dietary intake.

Cystilac seems to be practical, effective and convenient diet for CF children. It appears to be well tolerated and accepted by CF infants and children.

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211 Does an integrated clinical and nutritional approach prevent pre-diabetic decline?

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Pre-diabetic clinical decline is well documented in cystic fibrosis related diabetes (CFRD). We introduced screening for CFRD incorporating annual oral glucose tolerance testing in 1995. This is integral to an aggressive clinical and nutritional approach.

This case control study examines the impact of clinical approach on pre and post-CFRD clinical course. 48 CFRD patients (mean age 25.9±5.9 yrs) matched to 48 CF controls (25.9±6.3 yrs) for age, sex, Pseudomonas status had parameters of clinical status and nutritional intervention recorded annually from 6 yrs pre CFRD to 2 yrs post.

Weight and BMI increased throughout the study in both groups but were lower for CFRD at all time points to diagnosis (NS). Values were stable as % of control values [wt 96.6% (6 yr pre) v 97.5% (yr 0), BMI 98.9% (6 yr pre) v 98.0% (yr 0)] and were above controls 2 yr post diagnosis [wt 102%, 60.3 v 59.2 kg (NS), BMI 21.5 v 21 (NS)].

Lung function declined in both groups to diagnosis [absolute FEV1 63% to 46.5%, p=0.0008 (CFRD), 66% to 52.6% p=0.009 (CF)]. FEV1, FVC were below controls (NS), deviated from 1 year pre diagnosis [FEV1 as a % of controls = 95.4% (6 yr pre), 94.1% (1 yr pre) 86.6% (yr 0) NS], [FVC 96.2% (6 yr pre), 101.2% (1 yr pre) 93% (yr 0) NS] and stabilised on insulin therapy.

Nasogastric/gastrostomy feeding increased in both groups to diagnosis [18.8 to 43.8%, $\chi^2=6.9$, DF=1, p=0.008 (CFRD)], 8.3 to 18.8%, NS (CF)] deviating significantly from controls from 2 yrs pre diagnosis. IV treatment intensified, peaking at 1 yr post diagnosis 47.7 days/yr (CFRD) v 34.7 (CF) NS but there was no difference in nebulised antibiotic use.

An aggressive clinical approach prevents nutritional decline and delays respiratory decline until the year preceding diagnosis of CFRD.

212 Dietary treatment in adult patients with cystic fibrosis related diabetes

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Introduction: Cystic Fibrosis Related Diabetes mellitus (CFRD) is an increasingly recognized problem due to improved survival of patients with cystic fibrosis. Until now, dietary therapy has combined the conflicting principles of the usual dietary management of cystic fibrosis and diabetes mellitus.

Aim of the study: To investigate the need for new or adjusted recommendations in dietary treatment of adult patients with CFRD.

Methods: First, a literature research was performed, based on papers found in the databases Medline (from 1988 to the present), Cochrane, and PubMed, resulting in draft recommendations. Second, in November 2006 the members of the Dutch working group “Adult Cystic Fibrosis Specialist Dietitians” have discussed and amended these recommendations, providing a final guideline.

Results: Twenty three papers on detection and treatment of CFRD were found, but no evidence-based dietary guidelines. No meta-analyses or randomised controlled trials of dietary intervention in CFRD were found. As a consequence, recommendations are based on cohort studies and current clinical practice reported in clinical consensus guidelines. Adequate control of blood glucose appears to be no different from “normal” diabetes; energy recommendations however are higher than normal to maintain body weight; and saturated fats should not be restricted if body weight is too low because long-term effects on heart disease in this group are unknown.

Conclusion: A flow chart was developed to select different dietary treatments based on body mass index and abnormal plasma glucose and lipid levels. It can be used as a tool in the dietary treatment of the growing group of adult patients with CFRD.

213* Energy and nutrient intakes of paediatric subjects with cystic fibrosis (CF)

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Objectives: To estimate energy and nutrient intakes in CF paediatric subjects and to compare these intakes to Irish Recommended Dietary Allowances (RDAs; Food Safety Authority of Ireland (1999) and to CF dietary guidelines (Sinaasappel 2002, Borowitz 2002, NHS, UK, 2000) for children and adolescents.

Subjects: 100 CF subjects (53 boys, 47 girls), aged 9–19 yrs, (mean = 15.5 yrs) were recruited from a larger study being undertaken in Dublin, between Our Lady’s Children’s Hospital, Children’s University Hospital and the National Children’s Hospital.

Methods: Data on 3 day food records was collected coded and analysed using WISP (Tinuvial Software, UK). One-sample t-tests were used for comparisons and the independent-sample t-test was used to determine significant differences between boys and girls.

Results: There was no significant difference in energy and nutrient intakes between sexes. The CF recommendation of 120% for energy was achieved by 32% of the group. The RDA for calcium was achieved by 5% while 9% achieved the dietary vitamin D recommendation. Significant (p < 0.01) low intakes of iron were noted in 11–15 yr old girls. All subjects exceeded the RDA for vitamin C. Of the group 15% were receiving percutaneous endoscopic gastrostomy (PEG) feeds and 16% were consuming oral nutritional supplements (ONS). Intake of vitamin supplements was not assessed. A BMI of <50th percentile was evident in 69% of the group.

Conclusion: Particular emphasis should be given to the importance of energy and nutrient intakes for all CF children and adolescents. Early nutrition intervention is essential in promoting improved growth and lung function.